Section of Otology

President—R. R. SIMPSON, F.R.C.S.Ed.

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DISCUSSION: THE RAMSAY HUNT SYNDROME

INTRODUCTION

Mr. Kenneth Harrison, Department of Otolaryngology, Manchester University:

During recent years I have observed and treated a series of 12 cases of the Ramsay Hunt syndrome in Professor Victor Lambert's Department at the Manchester Royal Infirmary.

It was in Boston in June 1906 that Dr. Ramsay Hunt presented his classical communication to the American Neurological Association entitled "Herpetic Inflammation of the Geniculate Ganglion. A New Syndrome and Its Complications". He believed the syndrome to be dependent on a specific herpes zoster inflammation of the geniculate ganglion. This ganglion is situated on the facial nerve in the depths of the internal auditory meatus at the entrance to the bony fallopian canal (Fig. 1).

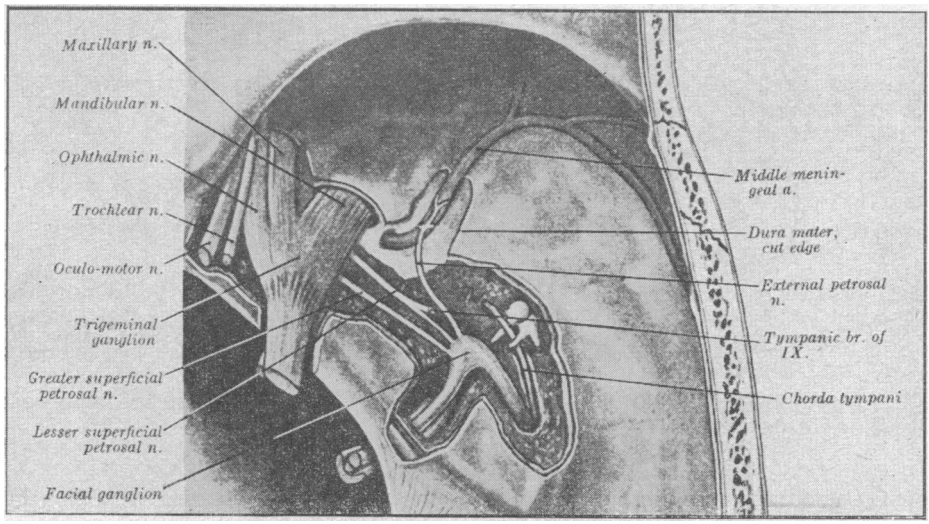

FIG. 1.—The geniculate ganglion of the facial nerve and its branches (reproduced from Gray's Anatomy by permission of Messrs. Longmans Green).

Classification.—He classified the different clinical types of the syndrome as follows:

(1) Herpes oticus, with no neurological signs.
(2) Herpes oticus with facial palsy.
(3) Herpes oticus with facial palsy and auditory symptoms.
(4) Herpes oticus with facial palsy with accompanying auditory and labyrinthine symptoms.

Each type is characterized by "pre-herpetic pains" localized to the ear and mastoid region. This is followed by a zoster eruption of variable extent, occurring in the area called by Ramsay Hunt the "zoster zone" for the geniculate ganglion (Fig. 2). It includes the tympanic membrane, external auditory canal and meatus, and the following areas on the lateral surface of the auricle, the concha, antitragus and the anti-helix and its fossa. Sometimes the eruption is seen on the postero-mesial surface of the auricle and the adjacent skin over the mastoid process.

The facial paralysis may occur soon after the appearance of the eruption, it is of the peripheral type and usually complete. Hunt considered that the motor fibres of the nerve were involved in the zoster inflammatory process.

Sometimes accompanying the foregoing symptoms there is loss of hearing and may be an associated tinnitus. He ascribed the auditory symptoms to an extension of the inflammation to the auditory nerve which is in close relationship to the geniculate ganglion.

He considered his fourth type to be a severe manifestation and along with the symptoms already mentioned were added those of vertigo.

He thought that the zoster inflammation extended along the nerve sheaths to the auditory nerve endings or *possibly* that the "ganglia" of the auditory nerve may be primarily involved in the zoster infection.

Hunt also emphasized a group of cases with zoster eruption and auditory symptoms *without* facial palsy. He believed this group may be due to a primary involvement of the "ganglia" of the auditory nerve, because he thought it improbable that an inflammation could extend to the auditory nerve without involving the motor fibres of the facial nerve.

He considered that the cutaneous sensory supply of the geniculate ganglion is an irregularly cone-shaped area, the apex corresponding to the tympanic membrane and the base situated on the external surface of the auricle; this is his so-called "zoster zone".

There may be "post-herpetic pain" which may persist for a considerable time after the zosteric eruption has disappeared.

It was in 1915 in his later investigation on the syndrome that he observed and described a zoster eruption sometimes occurring within the buccal cavity on the soft palate and the anterior two-thirds of the tongue (Fig. 3).

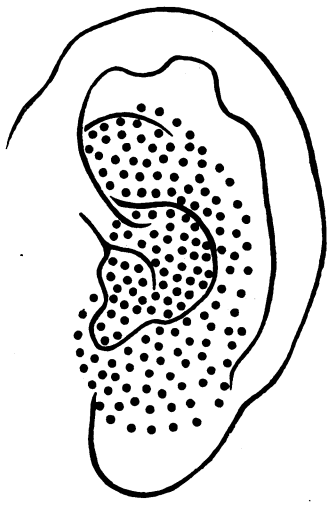

FIG. 2.—Distribution of vesicular rash over the "zoster zone" on the auricle (reproduced from *Brain*, **38**, 427, by permission of the Editor and of the publishers, Messrs. Macmillan & Co.).

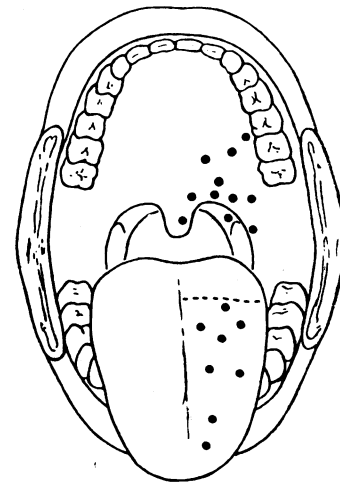

FIG. 3.—Distribution of vesicular rash over the soft palate and anterior two-thirds of the tongue (reproduced from *Brain*, **38**, 429, by permission of the Editor and of the publishers, Messrs. Macmillan & Co.).

Following this introduction of Ramsay Hunt's conception of the syndrome bearing his name, the clinical picture of the cases under review will be described.

CASES UNDER REVIEW

Since August 1949 12 cases of the syndrome have been observed and studied. It is thought best to assign the cases to the classification used by Ramsay Hunt. 8 of them were similar to his clinical group four, being herpes oticus with facial palsy and accompanying deafness and vertigo. 2 of the series were in his group three, herpes oticus with facial palsy and deafness. One of the remaining cases was a simple otic zoster with facial palsy, and the other a herpes facialis involving the mandibular division of the trigeminal nerve with an associated herpes oticus and facial palsy.

Occurrence.—All the cases have been seen between August 1949 and August 1953.

In August 1949 there were 4 cases, in May 1952, 2 cases, and in August 1953, 2 further cases; the other ones occurred singly and quite sporadically.

They all occurred between February and September of each year.

Age and sex incidence.—The patients were all adults between the ages of 33 and 76 years and included 10 females and 2 males.

CLINICAL FEATURES

The cases were all unilateral.

Usually there were prodromal symptoms of headache and lassitude with accompanying slight fever, the temperature being in the region of 99·5° to 100° F. These symptoms continue for a day or two.

Local symptoms then usually occur and it is during this stage that the patients are first seen. Firstly *pain* is complained of and it is of varying distribution and character. Earache is most frequently present and it is either felt superficially around the auricle or deeply within the meatus. Sometimes there may be generalized pain in the head but more frequently it is localized in the face and lower jaw and may radiate down the neck. It is often characterized by a burning or pricking sensation and there is an accompanying hyperæsthesia. At times the pain is actually neuralgic, and is referred to as pre-herpetic neuralgia.

Zosteric eruption.—Within a few days a zosteric eruption occurs. It is usually seen on the auricle and the external meatus, sometimes on the soft palate and occasionally on the anterior two-thirds of the tongue.

Eruption on the auricle and external meatus.—The skin of the conchal area of the auricle becomes reddened, swollen and tender and within a day or so vesicles are visible (Fig. 4).

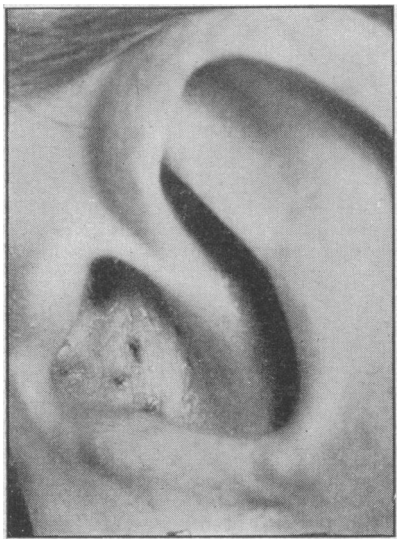

FIG. 4.—The vesicular rash confined to the conchal area of the auricle.

In our experience the vesicular rash has been confined mostly to the concha, anti-helix and its fossa and the external meatus. Vesicles on the drumhead have not been a common feature and no eruption has been seen on the postero-mesial surface of the auricle and the adjacent skin of the mastoid process. After the vesicles heal they may leave small scars.

In only one of the cases was there any facial eruption and this was widely distributed over the region supplied by the mandibular division of the trigeminal nerve. When the zoster eruption develops the pre-herpetic pain usually abates, but there is usually localized pain due to the eruption and accompanying adenitis.

Sometimes the walls of the external meatus may be so swollen that a clear view of the deeper parts of the canal is impossible.

Frequently the pre- and post-auricular lymph nodes are enlarged and tender and occasionally the upper deep cervical group.

In 2 cases perforation of the tympanic membrane was observed with accompanying inflammation of the meatal walls. At first it was puzzling as to whether the perforations were of recent origin due to the zoster infection or long-standing ones. In each case the patients denied previous aural disease, but after questioning and due consideration it was decided they were long-standing defects. In neither case did the perforation show any sign of healing.

Eruption of the soft palate and tongue.—In 5 of the patients small discrete vesicles were seen on the soft palate of the affected side and in 3 of these there were vesicles on the anterior two-thirds of the tongue with accompanying soreness. No noticeable pre-herpetic pain has been complained of within the buccal cavity, thus the vesicles will be missed unless particularly searched for.

Facial palsy (Figs. 5 and 6).—A lower motor neurone palsy was present in each case and usually of quite severe degree. There was always a latent period before the onset of the palsy and it was anything between one and ten days following the appearance of the vesicular rash. This latency may

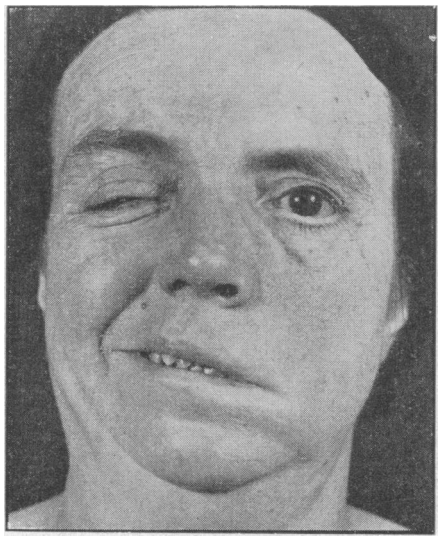

FIG. 5.—Complete facial palsy.

FIG. 6.—Wide distribution of vesicular rash on the auricle.

depend on the time it takes the inflammation to extend from the geniculate ganglion to the motor fibres.

Before the appearance of the palsy it may be quite difficult to make a correct diagnosis.

Paresis of the soft palate.—In none of the cases was a paresis of the soft palate noted. This finding was observed by Ramsay Hunt, and it has been reported occasionally in the literature.

Loss of taste.—In half the cases there was loss of taste over the anterior two-thirds of the tongue on the affected side. The taste sense always returned, usually within a period of a few weeks.

Deafness.—In 10 there was an accompanying perceptive deafness in the affected ear. Audiometric tests showed an increasing loss in the higher tones particularly at a frequency of 2048 and above (Fig. 7).

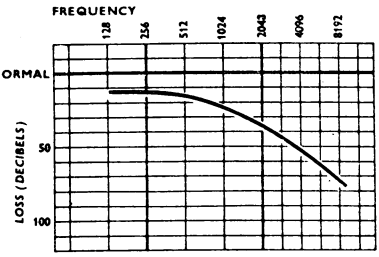

FIG. 7.—Pure-tone audiometric curve showing hearing loss in the higher tones particularly at a frequency of 2048 and above.

In 3 the deafness was bilateral and this was quite a surprising finding, the significance of it will be discussed later.

Tinnitus.—Was not a constant symptom even in the deafness cases and occurred in 4 of them. It was not unduly troublesome and always disappeared within a short time.

Vertigo.—Was present in 8 patients of the series and usually occurred within the first week of the infection. It was mostly severe with accompanying vomiting, and usually lasted from a few days to a number of weeks and necessitated rest in bed.

Even after the acute vertigo settled there was a feeling of unsteadiness, which in some cases lasted for many months, and in certain of the most severe ones the unsteadiness persisted for two to two and a half years.

In the early stages of the vertigo a spontaneous fine horizontal nystagmus was seen towards the sound side. Hot and cold caloric tests were carried out as soon as the patients were sufficiently fit and in two-thirds of the group with vertigo an absent response was noted on the affected side. The tests have been repeated at intervals since, sometimes as long as four years after the initial infection and no recovery has been noted.

The 3 patients with bilateral perceptive deafness had an accompanying vertigo and unilateral herpes and facial palsy. The results of the caloric test were interesting; there was an absent response on the side with the facial paralysis but quite a normal reaction on the other side.

Post-herpetic pain.—Post-herpetic pain has not been a troublesome symptom in any of the cases.

INVESTIGATIONS

(1) White cell count—no significant change noted.
(2) Audiometric and caloric tests—the results of these have already been described.
(3) Lumbar puncture.

In 5 of the cases lumbar puncture was performed. In 3 of them there was an increased mononuclear cell count ranging from 6–159 cells and a raised protein of 45–65 mg./100 ml. Within a few weeks the fluid was normal.

Greenfield and Carmichael (1925) state that a mononuclear pleocytosis may occur in herpes zoster. They do not consider there is any relationship between the severity of the disease and the degree of change in the cerebrospinal fluid. Lange (1908) described the facial and auditory nerves as having a common sheath formed by the continuations of the meninges. The auditory nerve at its entrance into the internal meatus is surrounded by a loosely adherent process of arachnoid until it reaches the fundus of the canal, at which point the membrane becomes adherent to the bony wall. This arachnoid covering follows the facial nerve for a short distance in the fallopian canal where it also becomes attached to the bony wall.

It is thought that because of these attachments of the arachnoid, the ganglia of the facial and auditory nerves are *intrameningeal*, and thus when involved in zoster infection changes may result in the cerebrospinal spinal fluid.

TREATMENT

There is no specific treatment for herpes zoster infection.

It is important to relieve the "pre-herpetic pain" and prevent infection of vesicular eruption.

The facial paralyses were treated by galvanism and, if possible, it is preferable to treat daily. It is

thought that galvanic stimulation aids in preventing further muscle wasting and delays fibrosis. It helps in maintaining the blood supply and when the nerve begins to conduct there is usually a better recovery.

Some surgeons practise decompression of the facial nerve in these cases and I am most interested to know if this does aid recovery.

In a number of cases, actually 6, a trial was made with the newer antibiotic drugs. Chloramphenicol was used in 3 and Aureomycin in the other 3. In only 1 case was there dramatic improvement.

It occurred in a female patient aged 33 who had the severe manifestation of the syndrome accompanied by symptoms of deafness and vertigo. Chloramphenicol was prescribed. Within a few weeks the perceptive deafness improved to within normal limits, the facial paralysis recovered and the vertigo disappeared. The caloric reaction has remained absent, even after a period of eighteen months.

The other cases showed no rapid improvement and progress was slow.

Prognosis

Post-herpetic pain has not been a troublesome symptom.

Recovery of facial palsy.—Only 10 of the cases can be traced. In half of them recovery was complete. In the other half 3 showed partial recovery *only* after a period of two years or more. It is too early as yet to know just how much recovery will take place in the other 2, as it is only three to four months since the palsy occurred.

Taste.—Always recovered and usually within a few weeks.

Deafness.—Only in half the cases of perceptive deafness was there improvement to within normal limits. In the 3 patients who presented with bilateral deafness, the pure-tone air conduction curves have remained about the same, 2 four years after the infection and the other three years.

Vertigo.—The vertigo has always disappeared sometimes taking many weeks, occasionally months. Even then certain patients have complained of unsteadiness which has lingered for as long as two to two and a half years.

Comment

The Ramsay Hunt syndrome appears to be quite rare, as only in 12 cases was the diagnosis made during a four-year period.

One-third of them occurred during the month of August 1949.

Two-thirds of the cases are placed in Ramsay Hunt's fourth group, in which there is accompanying deafness and vertigo, and he considered this group to be the severest manifestation of the infection.

It is thought that the zoster inflammation may extend from the geniculate ganglion of the facial to C.N. VIII via the communicating branches between the two nerves in the internal auditory meatus and at this level they are within a common sheath, or quite possibly there may be a primary involvement of the "ganglia" of C.N. VIII.

The 3 cases which were found on audiometric testing to have bilateral perceptive deafness were puzzling. There were no records of the patients' hearing before the infection, and they all denied any deafness. On the contralateral side in which there was deafness only, the spiral ganglion may have been *primarily* involved or there could have been a more central involvement. I have not found similar cases described in the literature, but Stewart (1927) reported a case in which there was a marked unilateral Ramsay Hunt syndrome involving C.N. VII and VIII and on the opposite side the vestibular ganglion *only* was thought to be involved, as evidenced by absent response to the cold caloric test.

In one of the cases there were additional small vesicles on the posterior pharyngeal wall and on the lateral border of the epiglottis and aryepiglottic fold; the movements of the vocal cords were quite normal. This type of case indicates involvement of C.N. VII, VIII, IX, and X. McKenzie (1915) reported a case of unilateral herpes zoster in which C.N. VII, VIII, IX, and X were involved. In his case the recurrent laryngeal nerve was implicated, too, resulting in a vocal cord palsy which recovered quite spontaneously. In 1943 Negus and Crabtree described a somewhat similar case in which there was involvement of C.N. VII, IX, and X. Along with marked vesicular eruptions, there were unilateral paralyses of the face, soft palate, constrictors and vocal cord; satisfactory recovery took place.

In none of these cases was the complement-fixation test performed to identify the zoster virus, as the facilities were not available. In 1933 Brain and Aitken carried out serological tests on cases of facial palsy and reached the following conclusions: A series of 9 cases of Bell's palsy *with* zoster-like eruptions of the auricle and meatus was found to contain antibodies to zoster virus as shown by a positive complement-fixation test. A series of 22 cases of Bell's palsy *without* zoster-like eruptions was found to have positive complement-fixation tests *in* 4 *cases* only. In these 4 cases it was thought that there may have been eruptions in the external auditory meatus which had been missed; thus it is always important to examine the meatus in the case of Bell's palsy.

Another point arising is that an eruption may be very slight or even missing, the so-called *herpes sine herpete*. If this is so, such cases would be classified as ordinary Bell's palsy unless there were signs of obvious involvement of C.N. VIII.

In none of the cases has any eruption been seen within the nasal cavity. This finding was reported in 2 cases of the Ramsay Hunt syndrome by Wakeley and Mulvaney in 1939; there was "slight serosanguineous nasal discharge" and small discrete vesicles were seen within the anterior part of the nostril on the affected side and also a vesicular rash on the hard and soft palate.

CONCLUSIONS

(1) The Ramsay Hunt syndrome appears to be an uncommon clinical entity, only 12 cases presenting during the four-year period from August 1949 to August 1953. One-third of the cases occurred during one particular month, August 1949.

(2) The sex incidence was predominantly female occurring in 10 of the series.

(3) The clinical picture of the syndrome is puzzling, making early diagnosis difficult. Antecedent otalgia may be misunderstood before the vesicular eruption appears. Even at this stage diagnosis can be problematical, and the buccal cavity should always be examined for the presence of vesicles on the palate and anterior two-thirds of the tongue.

(4) In this series a zoster infection involving the facial and auditory nerves was common, occurring in 10 out of the 12 cases. Involvement of C.N. IX and X appears to be very rare, occurring once only and there appear to be few reported cases in the literature.

(5) Complement-fixation test for zoster virus:

A positive test may be very helpful as confirmatory evidence of a zoster infection and particularly in cases of peripheral facial paralysis when no vesicular eruption can be seen.

(6) The complete recovery of the facial paralysis only occurred in half the cases traced.

(7) The perceptive deafness may be permanent as in 5 cases of the series, and sometimes is quite severe, but vertigo is never persistently troublesome.

(8) At present there does not appear to be any specific treatment for zoster infection.

I wish to thank my Chief, Professor Lambert, for allowing me to carry out this investigation and for his guidance and helpful criticism. My thanks are also due to Dr. R. G. W. Ollerenshaw and the staff of the Department of Medical Illustration at the Manchester Royal Infirmary.

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Dr. John D. Spillane:

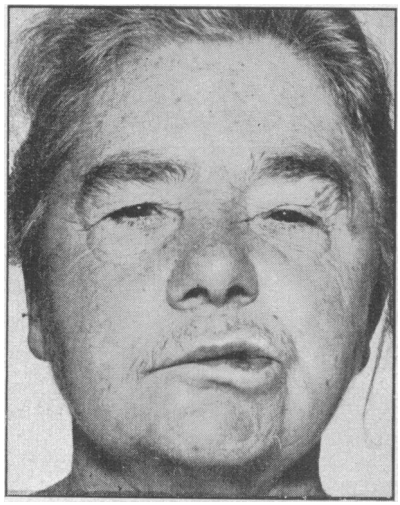
James Ramsay Hunt (1874–1937) was an American neurologist of Quaker Philadelphian heritage. He made definite contributions to neurological science. He described a form of akinetic epilepsy, certain cerebellar phenomena and occupational palsy of the deep palmar branch of the ulnar nerve. At one time or another three or four of his syndromes have been christened Hunt's disease. But his main interest throughout his life concerned the syndrome of "geniculate zoster". Indeed, his first paper on this subject was published in 1907 and the last was in the year of his death in 1937.

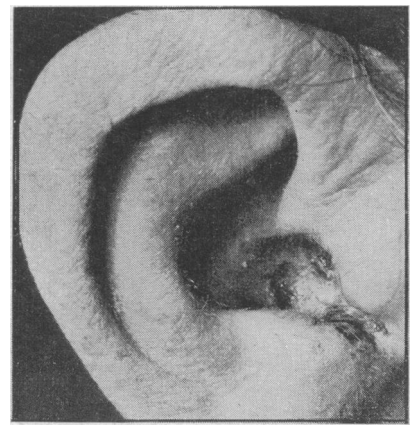
Ramsay Hunt looked on herpes zoster of the head and neck as due to involvement of ganglia containing the posterior spinal or unipolar type of cell. He observed, as others had done before him, that herpes oticus was sometimes combined with ipsilateral facial palsy. Some patients affected in this way suffered from tinnitus, deafness and vertigo. Hunt postulated that the geniculate ganglion, the homologue of the posterior spinal root ganglia, situated on the exclusively motor facial nerve, was in fact the site of infection in this type of case. He obtained no pathological proof of this theory but for over thirty years he defended and elaborated it with great, but in my view needless, ingenuity. He considered that swelling of the geniculate ganglion caused pressure on the facial nerve with resulting palsy. The auricular eruption was thought to indicate that there were in fact in this apparently motor facial nerve sensory fibres with cutaneous endings in a small area in the centre of the auricle which Hunt called the geniculate zone. When trigeminal or cervical herpes developed in association with facial palsy Hunt concluded that the gasserian or cervical ganglia as well as the geniculate ganglion were also affected. It was this conception of simultaneous ganglionitis which Hunt elaborated. But here he encountered difficulties. With trigeminal or cervical ganglion infection why should an associated facial palsy be attributed to involvement of the geniculate ganglion in the absence of herpes oticus? Was he to assume that this ganglion is infected and swollen so as to cause pressure on the motor fibres of the facial nerve and yet not manifest itself in the ordinary way by herpes in its alleged sensory zone?

Hunt was able to procure a post-mortem on one of his cases, a man of 48, who died eighty days after an herpetic infection causing facial palsy and characteristic eruption of occipito-cervical distribution. There was no auricular herpes. Unfortunately, the geniculate ganglion was never examined, but degeneration was found in the third cervical ganglion and its posterior root and in the nerve of Wrisberg between the geniculate ganglion and the pons. The post-mortem described in 1944 by

Denny-Brown and his colleagues was of a similar case who died sixty-four days after the onset of the illness. There were facial palsy and herpes of the second cervical distribution. It cannot be said with certainty that in this case there was an herpetic eruption in Hunt's "geniculate zone". The skin in the external auditory canal was "swollen, red and desquamating". The vesicular eruption was "in the right occipital and posterior auricular regions". Namely, in the second cervical dermatome. The photographs reveal only the facial palsy and the occipital herpes. At autopsy the geniculate ganglion was normal but the facial nerve was infiltrated throughout its course—above and distal to the geniculate ganglion. The second cervical ganglion was destroyed by the virus. The third cervical ganglion was practically intact. The ninth and tenth ganglia were not examined. This case demonstrates that in herpes of the head and neck damage to the facial nerve may occur without geniculate ganglionitis. It does not prove that geniculate ganglionitis does not occur and could not cause facial palsy and herpes oticus. Indeed, in view of the demonstration that herpetic infection of the relatively remote cervical ganglia can cause a neuritis of the facial nerve there can be no objection to the idea of geniculate infection doing likewise. But we now know that invasion and not compression of the facial nerve is the likely mechanism of involvement.

Confusion arises when the term Ramsay Hunt syndrome is applied to facial palsy with trigeminal or occipito-cervical herpes. There is no doubt that in such cases the anterior and posterior surfaces of the external ear respectively may be affected. The term should be restricted to those cases in which the herpes is confined to the concha and meatus (Figs. 1 and 2). If there is also herpes of glosso-





FIGS. 1 and 2.—Ramsay Hunt syndrome illustrating peripheral facial palsy with herpes zoster of cervical II and III distribution and with herpetic eruption in the conchal zone of the ear. There was no herpetic involvement in the mouth, pharynx or larynx. The facial paralysis developed twelve days after the first appearance of the herpetic rash on the neck. Facial paralysis may be a complication of herpes zoster in any zone of the head and neck, but is most commonly encountered in association with cervico-occipital zoster.

palato-pharyngo-laryngeal distribution then the IX and X ganglia are clearly implicated and otitic herpes could then be interpreted as occurring in the distribution of the auricular branch of the vagus nerve.

Sensory fibres to the concha from the facial nerve.—That meticulous observer Gowers wrote that in facial palsy: "I have several times found an area of anæsthesia on the front and back of the concha in the region of skin supplied by a nerve given off by the facial as it emerges." Gowers (1888) thought it may have derived from the V nerve. Larsell and Fenton (1928) demonstrated in man fibres from the pars intermedia of the facial nerve joining the auricular branch of the vagus nerve and being distributed to the skin of the auricle. Furlow (1942) stimulated the exposed pars intermedia of the facial nerve at open operation in a conscious patient and produced sharp meatal pain. Section of the nerve was not followed by any area of sensory loss but the meatal neuralgia ceased. Similar relief of meatal pain has been obtained in glossopharyngeal neuralgia after section of the glossopharyngeal nerve.

Can the method of topographic analysis of the facial palsy in the Ramsay Hunt syndrome determine whether the lesion is suprageniculate, geniculate, or infrageniculate? It is most unlikely to do so for several reasons. Firstly, the lesion in the facial nerve may be distributed throughout the course of the nerve in a patchy and uneven manner as in Denny-Brown's case. Secondly, the facial paralysis may be incomplete so that the absence of such signs as are used to determine the level of a lesion in the facial

nerve are then unreliable guides. Thirdly, the spread of infection or degeneration along axones may secondarily implicate certain functions such as tear secretion or taste sensation.

But if we assume the responsible lesion in the Ramsay Hunt syndrome is in the geniculate ganglion and completely involves the facial nerve, can this be in any way deduced clinically? Paralysis of the stapedius muscle and the lacrimal portion of the orbicularis oculi muscle and loss of sensation on the anterior two-thirds of the tongue would ensue, but hyperacusis (phonophobia), epiphora and taste loss could also obviously arise in an infrageniculate lesion (Fig. 3).

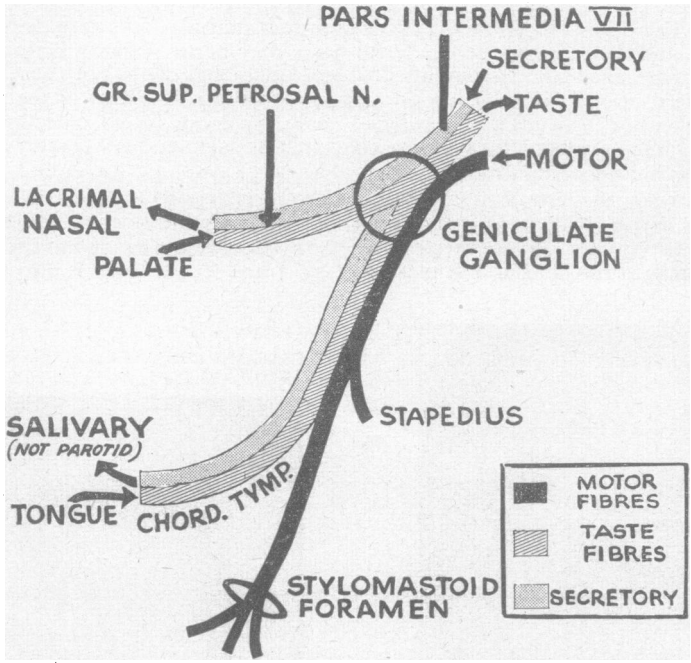


FIG. 3.—Course and designation of the components of the facial nerve.

The sensory root of the facial nerve, the pars intermedia, contains fibres transmitting the sensation of taste from the anterior two-thirds of the tongue and probably also from the palate. The former reach the geniculate ganglion via the chorda tympani nerve. Harris (1952) has effectively reasoned that taste fibres from the palate travel via the great superficial petrosal nerve to the geniculate ganglion. He thinks that there can now be little if any doubt that the gustatory fibres of the chorda tympani and the great superficial petrosal nerve pass centrally to the geniculate ganglion and thence by the pars intermedia to the medulla and pons. There has always been considerable divergence of view with regard to the suprageniculate pathway of taste fibres and even the direction of transmission of impulses in the great superficial petrosal nerve. Harris (1952) suggests that the presence or absence of loss of taste on the palate may be regarded as a clue to the position of the lesion in peripheral facial palsy. Suprageniculate or geniculate lesions should lead to loss of taste on the palate as well as on the tongue itself. However, I do not know of any observations on this point in the Ramsay Hunt syndrome. I have not found taste retained on the anterior two-thirds of the tongue on the side of the lesion in any case I have examined. Whether palatal gustatory sensibility proves a sufficiently reliable test I very much doubt.

Other sensory fibres in the facial nerve.—Secretory fibres to the salivary, lacrimal and nasal glands are present in the sensory root of the facial nerve above the geniculate ganglion. Those to the lacrimal and nasal glands leave the geniculate ganglion and travel in the great superficial petrosal nerve. Fibres destined for the submandibular and sublingual glands travel via the facial nerve and the chorda tympani. The parotid gland receives its secretory fibres from the auriculo-temporal branch of the mandibular nerve. Dryness of eye and nose, but not of the mouth, on the side of the lesion, may therefore indicate a geniculate *or* suprageniculate lesion.

Tschiassny (1946) used the involvement or otherwise of taste sensation and tear secretion to analyse the site of the facial nerve lesion in the Ramsay Hunt syndrome. His conclusions were that when facial palsy is associated with herpes of trigeminal or cervical distribution the lesion is below the geniculate ganglion. When the facial palsy is combined with auricular herpes as described by Ramsay Hunt, Tschiassny concluded that the lesion was at geniculate ganglion level. However, this analysis was

based on the supposition, which is probably incorrect, that taste sensation is not lost in a lesion of the nerve of Wrisberg—that is in a suprageniculate lesion. There is therefore no certain method of topographical analysis of facial palsy which will identify a lesion in the geniculate ganglion.

Pathology of herpes zoster.—The virus appears to spread from the meninges to the ganglia, thereby first affecting the peripheral ganglion cells. This explains why the eruption often begins in the median zone (Heilborn, 1950). It is abundantly clear that although the chief lesion is in the posterior root ganglion and is in the nature of a vascular disturbance (spinal ganglion apoplexy) lesions are found also in the posterior and anterior horn cells of the spinal cord, in the anterior and posterior nerve roots and the adjacent leptomeninges. There may even be encephalitis or myelitis. That is to say that the actual invasion of nervous tissues is more widespread than the eruption itself indicates. When there is motor paralysis associated with zoster in 90% of cases the paralysis and zoster are of the same segmental distribution (Taterka and O'Sullivan, 1943). The zoster usually precedes the paralysis by an interval of a few days or a few weeks. In the upper limb it is usually the deltoid and in the lower limb the quadriceps muscles which are affected. There thus seem to be factors of susceptibility and extension of infection by nervous pathways. In the case of zoster of the head and neck the facial nerve is the most vulnerable. Paralysis of the III, IV, V or VI cranial nerves is a rare occurrence. In trigeminal herpes facial nerve paralysis is commoner than trigeminal paralysis. The various clinical patterns are probably determined by factors of susceptibility and modes of spread of infection in an intricate neural network.

Selectivity of the zoster virus.—Lastly, there is the factor of selective choice of the zoster virus for certain cells within an individual ganglion. In Head's (1910) original account of 416 cases of herpes zoster there were 22 of trigeminal distribution: mandibular division 2, maxillary division 2, ophthalmic division 18. If the gasserian ganglion can be invaded in this fractional manner by the herpes virus we might expect it to take place in other ganglia—including the geniculate, the IX and X—with a consequent restriction of the eruption.

SUMMARY

(1) The facial nerve is the most vulnerable of the cranial nerves in zoster of the head and neck. Zoster infection is not confined to nerve ganglia. There is clinical and pathological evidence that it may cause neuritis of motor nerves. It is accordingly not necessary to postulate invasion of the geniculate ganglion when facial palsy is associated with herpes of the head or neck.

(2) Nevertheless there is no reason why zoster of the geniculate ganglion should not occur. Facial palsy would be a likely complication and the herpes, so far as is known, should appear in the conchal zone. No one has yet disproved Hunt's thesis that geniculate zoster is responsible for facial palsy with otitic herpes. The evidence indicates, however, that Hunt over-elaborated his thesis.

(3) The innervation of the external ear is complex, perhaps overlapping and variable. Aside from anterior and posterior auricular eruptions resulting from trigeminal and cervical ganglionitis, eruptions in the concha and meatus may result from VII, IX, and X ganglionitis.

(4) The geniculate level of a facial palsy cannot be identified clinically.

(5) The term "Ramsay Hunt syndrome" should be used only for those cases of facial palsy in which the herpes is restricted to the conchal zone. Only an autopsy in such a case can provide the solution to this problem of geniculate ganglionitis.

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Mr. J. P. Monkhouse: I should like to give a brief account of 3 cases which were admitted in October and November 1953 and January 1954, in order to point out the anomalies that occur in the Ramsay Hunt syndrome.

The first, a male aged 58, was admitted having had pain in the right ear for a week. He had a slight facial weakness, commencing herpes of the geniculate area but no vertigo or subjective deafness. An audiogram taken four days later showed a high tone loss on both sides, with masked bone conduction on the right following the same pattern, one which, at his age, might well have been present before his illness.

Five days after admission, the facial palsy was complete and the herpes very marked. On the sixth and thirteenth days, the facial muscles reacted normally to faradism and on the next day, the fourteenth, slight voluntary movement returned. In a month the face was fully recovered and there was a great improvement in hearing for high notes, both by air conduction and bone conduction, on the right. Three months later, his caloric reactions, which had not been tested before, were normal. He was treated with Chloromycetin for two days, followed by Aureomycin for five. No claim can be made on behalf of these drugs, particularly in view of the known variations in the course of this disease.

I want to emphasize the marked herpes, suggesting at the least a reasonably severe affection of the geniculate ganglion, coupled with a VII nerve that never shows the reaction of degeneration and recovers very quickly, a hearing loss that recovers and no evidence that the vestibular labyrinth was ever involved.

The next case was a male, aged 44. It is true that he never had any herpes, but on the other hand, he does not fit in with the usual conception of a Bell's palsy. Like the others, he had pain around the left ear for some days. Then there appeared a facial weakness and severe nausea, with constant vomiting and retching, which was accentuated by movement of the head. Two days later, the paralysis was complete and caloric tests showed a moderate left canal paresis. At ten days, the face began to move and, at the same time, an audiogram showed a left high tone loss by air conduction and bone conduction, with partial recruitment. At two months, the face and hearing had returned to normal. The left canal paresis was still present, and, to the same degree, at four months and, when tested again just recently, at twelve months.

In view of the severe vertigo, I thought I would try the effect of blocking the stellate ganglion, and it is interesting that except for one vomit while taking some tablets shortly after the injection, this symptom ceased abruptly. I had also heard at that time of an intravenous procaine drip as a treatment for facial palsy and this was given over the fifth and sixth days. Again it is impossible to assess the value of these procedures.

Here I want to stress, no herpes, complete facial palsy and objective involvement of both portions of the labyrinth. There was never R.D., the face began to recover in ten days and was well in two months. The hearing returned to normal, but the canal paresis persists.

The last case was a female, aged 32. Again there was pain, together with tinnitus in the right ear and a slight vertigo for four days before the onset of facial palsy. Two days later the palsy was complete, there was severe vertigo and vomiting, marked herpes of the geniculate region, loss of taste, no subjective deafness but she complained of hyperacusis. The mouth and palate were normal.

Denny-Brown *et al.* (1944) showed in one particular case in which a facial palsy was present that the geniculate ganglion was not affected and could not have been the cause of the palsy. They found a poliomyelitis and a neuritis of the facial nerve and put these forward as an alternative explanation of the Ramsay Hunt syndrome.

During a review of recorded cases, they would appear to admit the possibility of variation in the pathology when they say, "Nor is it certain that herpes zoster does not on occasion affect the geniculate ganglion", but they finish the article with the uncompromising statement, "Analysis reveals that the evidence for geniculate ganglionitis in the Ramsay Hunt syndrome is invalid".

My patient now came forward with a most interesting observation. She had caught a cold but said that it was only on one side. On the left side, her nose was hot, stuffy and running, while on the right, the side of her facial palsy, she just had not got a cold. It was evident that the secretory fibres of the parasympathetic were out of action and it seemed likely that the vasodilator fibres would be similarly affected, with the result that the nasal temperature should differ between the two sides. Dr. J. C. Seymour and Mr. J. W. Tappin, of the Ferens Institute of the Middlesex Hospital Medical School, made recordings, and I should like to thank them for a very great deal of help. Using a small thermocouple, connected to a large amount of electrical apparatus presided over by Mr. Tappin, it was possible to take instantaneous temperature readings.

Fig. 1 gives the dates on which the clinical signs and symptoms were first noted, together with their subsequent course, and the figures obtained during the investigation of the nasal temperatures. The sense of taste on the anterior two-thirds of the tongue was lost from the beginning and has not recovered. The secretion of tears was markedly diminished for two and a half months, when recovery commenced, and this was followed by a period of hypersecretion, suggesting that the recovering nerve was over-acting. The same phenomenon was observed in regard to nasal secretion.

Lacrimal secretion was tested by Schirmer's method and it must be emphasized that an objective test is essential. The drooping of the lower eyelid associated with facial palsy removes the lacrimal punctum from contact with the globe and results in failure of the mechanism of drainage, so that the eye may appear to be swimming in tears when, in fact, secretion is minimal. It was not realized that the nose was involved until the patient caught a cold and noted the absence of secretion on the affected side. The entry "equal" on March 20 only means that the cold had got better and it is not till a week later, at two months, when the right side begins to over-secrete, that we can be sure that recovery has commenced. This eventually settled down and in October, when the patient caught another cold, both sides of the nose reacted in the usual manner.

The figures for nasal temperatures, though they conform to theoretical expectation, are offered with diffidence since this investigation has only been carried out on one case, and it will be repeated if further material becomes available.

1953	Taste	Tears	Nasal Secretion R.	Nasal Secretion L.	Intr.Nasal Temp. R.	Intr.Nasal Temp. L.	Temp.after Pilocarpine R.	Temp.after Pilocarpine L.	Temp.after Benzedrine R.	Temp.after Benzedrine L.	Facial Nerve	
31 Jan.	Absent										Complete palsy	Herpes
8 Feb.		Eye dry										
10 Feb.											R.D.	
13 Feb.		Greatly reduced										
17 Feb.											R.D.	
19 Feb.	Absent											Herpes gone
22 Feb.		Almost nil										
24 Feb.				+								Cold
6 March	Absent	Dimin.		+	34	36	34	36	33	33	Complete	
20 March	Absent	Dimin.	Equal		34.5	35	37	36.5			"	
27 March			+									2 mths.
10 April	Absent	Equal	+		35	34			32.5	33.5	"	2½ mths.
8 May	Absent	Slight+	+		35.5	34.5					"	
13 May											Slight movement	3½ mths.
14 June			Equal									
6 July	Absent	Slight+									Marked improvement	
21 Oct.			+	+								Cold
19 Nov.	Absent	Equal									Good but not normal	10 mths.

FIG. 1.

During the period of parasympathetic paralysis, the resting temperature on the right side was lower than on the normal side but when recovery and over-action had occurred the temperature became raised above the normal. The readings obtained after stimulation of the parasympathetic with pilocarpine and of the sympathetic with Benzedrine fall into place if one can assume that a blood vessel is only capable of a certain maximum contraction or dilatation and that, normally, it lies in a mid-condition, being influenced simultaneously in both directions. If, therefore, a vessel which is deprived of its dilator fibres and consequently is near to a state of contraction, is subjected to a dilator stimulus, the effect will be greater than would result from the same stimulus applied to a normal vessel which is nearer to a state of dilatation. On the first occasion, the dose of pilocarpine was small and produced no change on either side, but with a larger dose there was a rise of temperature of 2·5° C. on the abnormal side but only of 1·5° C. on the normal. Benzedrine, producing the opposite effect, is only able to lower the temperature by one degree on the abnormal side where the vessels are already near to full contraction, but lowers the temperature by 3 degrees on the other side. On April 10, when Benzedrine was used for the second time, there is a change in the situation in that the parasympathetic has recovered and is, in fact, over-acting. Now the vessels on the right side are abnormally dilated and react more to a constricting stimulus than do those on the normal side, with the result that temperatures drop 2·5° C. on the right as against 0·5° C. on the left.

I would therefore suggest that in this particular case we must, unless we are prepared to assume the presence of multiple and widely spaced lesions, agree that the ganglion is at fault. Further confirmation is afforded by the order of return of three of the lost functions and the failure of return of the fourth. Taste fibres have their cells in the ganglion, but with the eye, nose and face it is only a matter of fibres passing through or around the ganglion. The secretory fibres to the eye and nose are of much smaller diameter than are the motor fibres of the facial nerve and it is known that small diameter fibres withstand pressure better than do large ones. This all fits in: the nose and eye recover first, in two and two and a half months respectively, the face takes longer, three and a half months, while the taste has not recovered in ten months, and if the cells are destroyed, it never will. There is still the problem of the VIII nerve, which is affected in a curiously selective manner.

The audiograms in Fig. 2 show the complete recovery of hearing, a feature common to all 3 cases, but the vestibular labyrinth behaves very differently. The damage was marked from the first, but after six months it became worse, so that now there is no reaction even to water at 20° C. In the second case, the vestibular damage, though persisting, has not increased.

Shute (1951) describes the facio-cochlear anastomosis as a connexion between the cochlear nerve and the ganglion itself. There is Oorts bundle constituting a pathway between the cochlear nerve and

the inferior vestibular ganglion, and, lastly, the three portions of the nerve and the vestibular ganglia are all closely applied to each other. Since it is known that a neurotropic virus can track in both directions along either sensory or motor nerves, it seems reasonable to accept the possibility of a direct spread to some or all portions of the VIII nerve. It is not, however, so easy to explain the differing fates of the cochlear and vestibular labyrinths. The recovery of hearing must imply that the spiral ganglion escapes and, conversely, the permanent damage to the vestibular apparatus suggests that the vestibular ganglia are involved. It would be most attractive to conclude that the deafness is a secondary and temporary phenomenon resulting from pressure of the swollen ganglia upon the cochlear nerve and capable of recovery. Unfortunately, my first case deals this theory a severe blow

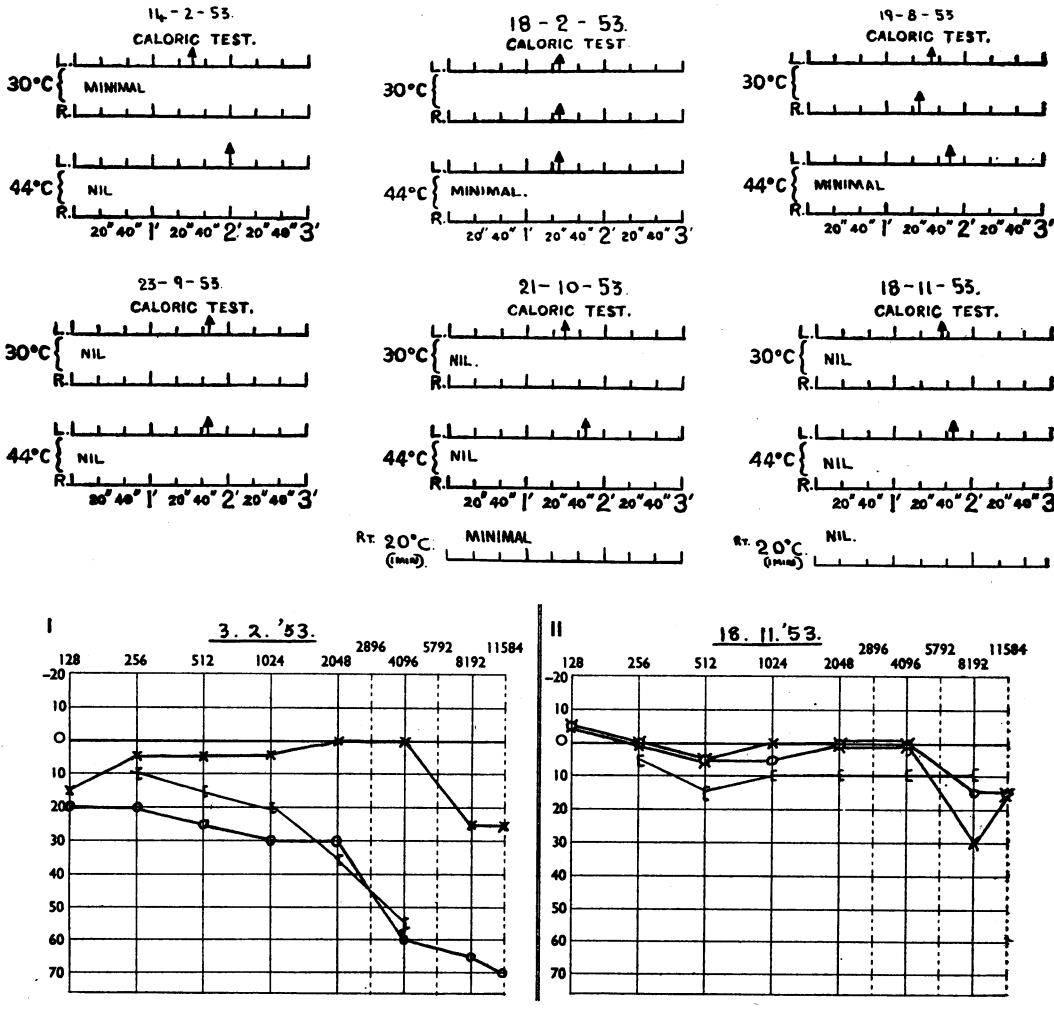

FIG. 2 (*Case III*).

in that, although deafness was present, we did not, admittedly with an incomplete investigation, find any evidence of vestibular damage. Alternatively, it could be held that the lesion of the cochlear division of the nerve is limited to a neuritis which can recover but that, should the infection spread to the vestibular portion, there ensues an additional ganglionitis, which is irreversible. Some virus infections are known to be very patchy in their distribution, but it is difficult to believe that the infection would always die out before reaching the spiral ganglion, and I must own that I am not satisfied with either of these explanations.

These cases present curious anomalies. Only 2 have herpes, but all have a facial palsy. The severity of the palsy has no relation to the extent of the herpes. 2 have permanent damage to the labyrinth

but the third escapes. In fact the only symptom common to all is a deafness that recovers. It would seem that the site or sites of the lesions in this disease are very variable and I feel that it would be no more justifiable to assume that Ramsay Hunt was entirely wrong than it would be to claim that every case can be explained solely on his hypothesis.

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Dr. Wilfred Harris thanked the openers for their scholarly addresses. It was mentioned that some of the cases seen showed symptoms of loss of taste on the tongue and palate which was symptomatic of a case of geniculate neuritis. It was an uncommon syndrome and he had seen it in about 8 cases. Dr. Spillane rightly drew attention to the fact that the diagnosis of geniculate neuritis was not based on post-mortem facts, but the syndrome was so definite that he thought one was not wrong in continuing to call it geniculate neuritis until one knew better.

The first case he saw was over forty years ago at St. Mary's Hospital. There was severe pain in the ear with a bloody discharge from the meatus, and Dr. William Hill, otologist, was not sure of the cause. The hearing apparently was not involved. Dr. Hill syringed out the ear and the result was most agonizing pain lasting for weeks. When he examined the patient himself he found he had a typical rash of herpes on the posterior wall of the meatus and at the junction of the pinna with the scalp, and the actual tympanic membrane was also involved. There was no facial palsy and in process of time the patient recovered.

In another case with a similar syndrome loss of taste was definite. A colleague had told him that he had difficulty in testing a case of taste loss on the palate. The best way was to use an illuminated spatula to hold down the tongue and gently rub a moist swab soaked in a bitter solution on the palate. A good method is to use a weak galvanic current, touch the palate with one electrode when a sensation of a coppery taste will be noticed only on the normal side touched by the current. He had used that many times and was able to demonstrate the loss of taste on the palate in cases of facial palsy before classes of students and medical registrars. There was no question about the loss of taste on the tongue in cases of facial palsy. There was not such a large distribution of taste in the palate as on the tongue and in old people the test on the palate was not very satisfactory.

In 1909 or 1910 at a neurological meeting he mentioned that loss of taste was commonplace in facial palsy and he was jeered at by the Neurological Society members; he could not then understand why because he thought everybody knew that loss of taste on the tongue was common in facial palsy and now it was universally accepted.

Lesions in the geniculate ganglion might or might not be associated with facial palsy. The association might be accidental and muscular palsy applied also to herpes elsewhere in the body. Every now and again one saw it in the abdominal muscles. He had seen a total Erb's palsy of the biceps and deltoid and other muscles associated with a typical distribution of fifth cervical herpetic scarring.

Hunt having carefully described the syndrome in the early 1900s, the speaker remembered that Hunt was associated with a case of Pierce Clark's of tic in the facial region when the man had been suffering violent pain for years in this area. An operation was performed to divide the pars intermedia Wrisbergi and the case was cured. It was a marvellous opportunity for testing the taste function and he wrote and asked Ramsay Hunt what was the effect on taste and he replied that he was ashamed to say that neither he nor anyone else had thought of testing taste.

Herpetic pain could continue for years, it was an agonizing symptom and he knew of the case of a man suffering from shingles followed by severe long-continued herpetic pain; all sorts of treatments were tried, and somebody with more wit than brain suggested cutting off the whole area of skin affected by the scarring of the herpes. The man consented to this being done, he bore the pain of the incision without any anæsthetic, but the post-herpetic pain continued, and the man shot himself.

Mr. Norman A. Punt wished to report briefly 2 cases which suggested that the herpes zoster virus might affect various neural pathways at random, and that clinical estimation of its routes of spread and distribution should be accepted with reserve.

A man presented with complete left facial paralysis and diminished taste sensation; there were herpes vesicles over the postero-superior part of the tympanic membrane, meatus and neighbouring pinna. There were also a left palatal paresis, recurrent nerve paresis and possibly cricopharyngeus paresis. There were no mucosal vesicles. He had only transient deafness, due to the skin lesions.

A woman complained of pain in the right ear; the skin behind the ear was a little reddened, but there was no other ear lesion and no deafness. She developed a complete right facial paralysis with diminished taste sensation. Injection and herpes vesicles appeared and were confined to the right side

of the soft palate and posterior pharyngeal wall. There was a Horner's syndrome, and also nausea and giddiness which were believed to be vagal in origin, no cochlear or labyrinthine dysfunction being demonstrable.

Mr. A. Laskiewicz said that the topography and vascularization of the fallopian canal could be proved by the following method: a very thin metal wire, or any strong contrast medium, should be introduced into the lower end of the fallopian canal on the dried temporal bone. A radiograph taken in the Stenvers position and the lateral oblique position showed the course of the named canal which could be distinguished as four principal types: (1) the nearly right angular form about 75%; (2) the arched regular form nearly 25%; (3) the wavy form, 3%; and (4) the double arched form, 1% in which the point of junction of both arches corresponded nearly to the region of the geniculate ganglion.

The vascularization of the fallopian canal and the facial nerve was given by the stylomastoid artery, the stem of which was surrounded by the auricular branch of the vagus nerve and in cases of aneurysm of the lower part of this artery there was often complete anæsthesia of this branch. Secondly, small branches of the arteria meningea media and auditiva interna dealt also with the blood supply of this canal. The veins flowing off consisted of a formation of a fine venous plexus joined closely with the surrounding bony vessels. The stream was directed mainly towards the petrosal superior sinus and the vena auditiva interna.

In 2 cases of facial nerve palsy with herpes zoster cephalicus in men aged 41 and 46, the main complaints were of giddiness with hardness of hearing, burning pains in the margins and apex of the tongue, and unilateral facial nerve palsy. In both cases herpetical blisters were very evident in front of the auricle and around the entry to the external meatus; in the first case also on the right side of the palate. In both cases one was dealing with a considerable decrease of the sense of smell and parosmia, e.g. the smell of eau-de-Cologne was taken as the smell of oil or petroleum.

As to the sense of taste, there was parageusia, with permanent salty or bitter taste, unilateral hypogeusia for sweet or sour substances, besides a lowering of the feeling sensation on the given side. After administration of chloramphenicol 0·5 gramme three times daily for four days, supported by injections of vitamin B_1, these disturbances disappeared within three weeks, and only unilateral hardness of hearing remained.

Mr. Terence Cawthorne said that a matter of prime importance in dealing with cases of facial paralysis, was a full exploration of the various functions of the facial nerve. Besides enquiring into the state of the facial musculature it was necessary to test the ability to taste and to lacrimate. Hearing was also often mentioned because it was thought that paralysis of the stapedius muscle gave rise to sensitivity to loud low-pitched sounds. He had rarely noted this though he thought that at times it might occur. The reason why it was so rare was that the protective function of the stapedius muscle when it was paralysed was taken over by the tensor tympani which was supplied by the V nerve.

He had found that it was always advisable to test lacrimation. There were certain cases which superficially were like an ordinary Bell's palsy but in which lacrimation was impaired. There might also be involvement of the VIII nerve as well but no herpes. At one time he had thought that impairment of lacrimal secretion always meant a dry eye but this was not so, because the eye could be kept moist by secretion from other glands in the conjunctiva.

He had operated on one case of herpes facialis in which the face remained paralysed a year after the disease appeared. He exposed the nerve right forward to the geniculate ganglion and it was observed that the nerve in the neighbourhood of the geniculate ganglion was discoloured and swollen. Within two months of the operation there was a partial return of facial movements.